

Welcome to the Boston Children's Hospital Transition Toolkit!

As a teenager getting ready for adulthood, or as a young adult, you can begin to take control of how you handle your metabolic condition and your health in general. This Toolkit is designed to help you! To start, read through these materials and fill out the forms. You can also begin to look for an adult-focused doctor to replace your pediatrician who only deals with childhood problems. This process of switching to an adult-focused doctor is called *medical care transition* and takes time and planning – but once it's done, it will be great for your future health and success.

Use the following forms to help achieve your medical care transition



**Health Readiness
Assessment**



**Metabolic
Condition Basics**



**Medical Health
Summary**



Transition Plan

What am I supposed to do with these **forms**?

Measure your health independence by taking the [Health Readiness Assessment](#) and over time try to answer yes to every question.

Read up on your [Metabolic Condition Basics](#) and discuss possible adult-health problems with your doctors or nurses.

Ask a doctor or social worker for help filling out the [Medical Health Summary](#) and [Transition Plan](#) at your next appointment.

Save these forms on your computer and USB flash drive and bring the Transition Toolkit to every medical appointment.

Keep the flash drive in your wallet or purse and save any changes or updates.



Save these forms and bring them to future medical appointments



Ask a doctor to help you fill out your forms

How will The **Transition Toolkit** help me? With the Toolkit you can –

Decide when you're ready to act as your own health spokesperson.

Prepare for doctor's appointments and fill out health forms.

Talk to doctors and nurses about your medical condition and what roles they should play in your medical care.

Keep track of all of your important medical and condition information.

To get started, fill out the [Health Readiness](#) form.

Transition Toolkit

Health Readiness Assessment

Are You *Ready* to Handle Your Own Health Care as an Independent Adult?



To know if you are ready for a health care transition, answer the following questions:

1. I know my height, weight, birth date, and social security number Yes No
2. I know the name of my condition Yes No
3. I know my genotype or genetic status Yes No
4. I can tell you about my condition and explain my health care needs Yes No
5. I know what kind of medical insurance I have Yes No
6. I know where to find my medical records Yes No
7. I know who to ask about getting my own health insurance Yes No
8. I know the basic treatment for my condition Yes No
9. I know what my diet should include and not include Yes No
10. I know how to prepare my own food Yes No
11. I know how to shop for my own food Yes No
12. I know the names of my supplements and medicines Yes No
13. I know how to order or get my medications filled Yes No
14. I know the symptoms and medical problems related to my condition Yes No
15. I visit my metabolic specialist regularly Yes No
16. I have discussed issues about sex with my doctor Yes No
17. I know how to get birth control and how to stay protected against sexually transmitted diseases/infections (STDs) Yes No
18. I have discussed the use of tobacco, alcohol and drugs with my doctor Yes No
19. I know how to schedule a doctor's appointment Yes No
20. I keep a schedule of medical appointments on my calendar Yes No
21. I can get myself to medical appointments (or can arrange transportation myself) Yes No
22. I know who to call in case of an emergency Yes No
23. I prepare questions for my health care provider before appointments Yes No
24. I have a support network for my condition Yes No

Transition Toolkit

Health Readiness Assessment

Are You *Ready* to Handle Your Own Health Care as an Independent Adult?



Now count the number of times you checked YES.

If you checked YES for:

17 to 24 statements – great work!

You are already taking full responsibility for your health care. The transition forms in this Toolkit will help you become a stronger health advocate.

8 to 16 statements – you are on the right path!

You are actively taking on many important responsibilities in your health care. The following Transition Toolkit forms will help you answer YES to many more of these questions.

0 to 7 statements – let's get started!

Transitioning health care to adult providers and becoming a better health advocate for yourself takes time and planning. Now's a perfect time to start! Pick a few of the previous responsibilities from the list and spend the next month doing them. The Toolkit forms will make this much easier.

Now what?

Update and save this assessment every month until you've answered all of the tasks with a YES!

Next, take a look at some [basic information about your metabolic condition](#).



Overview of the Condition:

Phenylketonuria (PKU) is a rare condition in which your body cannot break down an important amino acid called phenylalanine (Phe), which is found in all protein foods. Because you can't break down this chemical, it builds up in your body and can cause serious health problems.

Possible Medical Problems for Babies and Children:

- All newborn babies are tested for PKU in the US.
- Babies with PKU who do not have good treatment while growing up tend to have worse PKU-related problems later in life.
- If a Phe-restricted diet is followed, babies will develop normally – if not, babies are at risk for mental retardation, delayed social skills, hyperactivity, seizures, skin rashes, and other medical problems.

Possible Medical Problems & Symptoms for Teens and Young Adults:

- Depression
- Anxiety and Panic Attacks
- Memory loss
- Attention difficulties
- Aggressive or angry outbursts
- Migraines
- Eczema
- Sleep problems
- Inability to finish reading a book
- Troubles in social relationships
- Planning, organization and other executive functioning skills
- Breath and skin may smell “musty”

How to Avoid Medical Problems and Complications:

- In severe forms of PKU, follow a strict low-phenylalanine (low-protein) diet
- Foods to avoid include: all dairy products (products with milk in them), meat, fish, eggs, nuts, regular pastas, regular rice, and artificial sweeteners like NutraSweet (aspartame) commonly found in diet soda.
- Your diet must also include special formulas and specially prepared low-protein foods, which supply protein building blocks and other nutrients needed to keep you healthy.
- Your diet must be monitored by a dietician who is familiar with PKU.

New treatments for PKU:

- **BH4 therapy:** This is a medication that boosts the activity of the enzyme that is not working in PKU. People with milder forms of PKU can lower their blood phenylalanine (Phe) levels and can sometimes eat more protein. Some people with classic PKU also benefit from this medicine.
- **Large Neutral Amino Acid therapy:** This is also a medication that competes with Phe transport across the blood-brain barrier. This means that less Phe enters the brain. Note: This medication cannot replace diet during pregnancy.

Transition Toolkit

Metabolic Condition Basics: Phenylketonuria (PKU)

Medical information you need to know as an adult with PKU



- Formulas: There are many new formulas (medical foods), some of which taste much better than the old formulas.
- Enzyme replacement therapy: This therapy is in clinical trials. This means that it is not yet approved for general use. Its safety and effectiveness are being checked

Ask your metabolic doctor or dietician for more information about these advances in therapies for PKU.

Fertility and Pregnancy:

- Women with PKU can have healthy pregnancies and deliver healthy babies if they maintain a strict low Phe diet and take the formula *before* and *during* pregnancy.
- Staying on diet both before becoming pregnant and throughout the pregnancy can prevent developmental problems and mental retardation in their babies.
- Pregnancies should be handled normally but the mother should be monitored by a doctor very closely
- PKU is passed on to babies only when both parents carry a copy of the gene
- This **Maternal PKU & Pregnancy** guide is for women with PKU who are planning a pregnancy or are pregnant: <http://newenglandconsortium.org/for-families/phenylketonuria-pku/maternal-pku-pregnancy/>

How To Get Support:

- Get education support from teachers and specialists at your school.
- Join a support group with people who have PKU. <http://www.pkunews.org/support/groups.htm>
- A lot of research is being done on PKU. Get up-to-date information about new treatments and discoveries from the The National PKU Alliance: <http://npkua.org/>

Resources:

Genetics Home Reference: <http://ghr.nlm.nih.gov/condition=phenylketonuria>

PKU Toolkit: <http://newenglandconsortium.org/toolkit/>

PKU.com: <http://www.pku.com/>

National PKU News: <http://www.pkunews.org/index.htm>

The National PKU Alliance: <http://npkua.org/>

Transition Toolkit (New England Consortium of Metabolic Programs)
<http://newenglandconsortium.org/for-families/transition-toolkit/>

Got Transition
<http://gottransition.org/>

Genetics Referrals:

Clinical Services
<http://www.ncbi.nlm.nih.gov/sites/genetests/clinic?db=genetests>

Genetic Services
<http://www.acmg.net/gis>

Transition Toolkit

Metabolic Condition Basics: Phenylketonuria (PKU)

Medical information you need to know as an adult with PKU



What Next?

Now you can fill out the [Medical Health Summary](#), print it, and save it. This will help you keep important medical information in one place.

Transition Toolkit
Medical Health Summary

Use this form to save important health information



Date Updated: _____

Last Name		First Name	
Birthdate	Gender	Weight	Height
Address			
City		State	Zip Code
Insurance Name			
Insurance ID & Group #			
Insurance Address & Phone Number			
Metabolic or Genetic Condition			
Medications/Supplements and Dosages			
Allergies to Food, Drugs and Other Substances			

Emergency Contact Name & Relationship		Emergency Contact Phone Number
Pharmacy Name		Pharmacy Phone Number
Immunizations:	Type	Date



Transition Toolkit
Medical Health Summary

Use this form to save important health information



Other medical information:

What next?

A final step toward completing your medical transition is meeting with your health care providers to discuss and fill out your [Transition Plan](#).



Transition Toolkit

Transition Plan

Make an ongoing health plan with your health care provider



Fill out this plan with your health care provider, to help you transition to being in charge of your own health

Patient Name: _____

Boston Children's Hospital Medical Record #: _____

Primary Care Physician	Phone Number
	Email
Pediatrician	Phone Number
	Email
Metabolic Specialist	Phone Number
	Email
Other Health Care Provider	Phone Number
	Email
Emergency Contact Name & Relationship	Phone Number

Medical Findings

Genotype/Laboratory Findings
Treatment Status
Current Medications/Treatments
Allergies/Adverse Reactions
Past Medical Concerns or Hospitalizations
Living Situation/Employment
Mental Health Status/Cognitive Status



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Transition Plan

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Other Health Risks

Contract:

To enhance collaboration, each person should agree on the following responsibilities:

Metabolic Specialist will:

- Obtain labs and tests: For example...
- Prescribe condition-related medications:
- Work with PCP to develop an emergency protocol:

Patient will:

- Remember to take medications: For example...
- Order and pick up supplies/supplements:
- Show-up to appointments:

Primary Care Physician will:

- Monitor patient's general health:
- Update metabolic specialist with important patient information:
- Contact metabolic specialist with medical concerns: For example...
- Other:

When you're done filling out this Transition Plan, print it and save it in a safe place.

Congratulations! You are well on your way to being in charge of your own health.